

The natural history of hidradenitis suppurativa

JM von der Werth,* HC Williams

Department of Dermatology, Queen's Medical Centre, Nottingham NG7 2UH, UK. *Corresponding author, Lincoln County Hospital, Greetwell Road, Lincoln LN2 5QY, UK, tel. +44 1522 512512; fax +44 1522 573101; E-mail: jan@vonderwerth.freereserve.co.uk

ABSTRACT

Aim To investigate aspects of the natural history of hidradenitis.

Background The natural history of hidradenitis suppurativa (HS) is not well known. There is incomplete published data on the average age of disease onset, progression of the disease, average monthly incidence and duration of boils, and factors that relieve or exacerbate disease symptoms.

Study design Questionnaire-based survey among HS patients identified from hospital records of three hospitals in Nottinghamshire, UK.

Results One hundred and ten of 156 questionnaires (70.5%) were returned, 93 from females and 17 from males. The average patient's age was 40.1 years and the average reported age of disease onset was 21.8 years. At the time of the survey patients had suffered an average disease duration of 18.8 years. Most patients (98 of 110) still had experienced active disease within the past year. There was some evidence that in women the condition has a tendency to ease or subside after the menopause. Forty-four per cent of women felt that their condition was aggravated by menstruation. Thirty-eight per cent of patients gave a positive family history of the disorder. The average duration of painful boils was 6.9 days. In addition, 62% of patients acknowledged the presence of permanently painful boils that failed to subside. Patients developed a median of two boils per month. Factors that could aggravate the condition were primarily sweating or heat, stress or fatigue and tight clothing or friction. Factors that could improve the condition consisted largely of a variety of medical treatments and a number of life-style measures, such as swimming or baths. Twenty-four per cent of patients had failed to find anything at all to help their condition, despite an average disease duration of almost 19 years.

Conclusions The study highlights several of the factors that make HS one of the most distressing dermatological diseases, such as the average monthly incidence of painful lesions, their average duration and the chronicity of the disease. It seems striking that the mean duration of an HS boil (6.9 days) roughly equals the duration of an average course of antibiotics. The postulated response of HS to oral antibiotics may thus simply have its explanation in the natural history of the condition itself.

Key words: hidradenitis suppurativa, natural history, treatment

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Introduction

The first description of hidradenitis suppurativa (HS) is credited to the French Physician Velpeau. His countryman Verneuil termed the disease 'hydrosadenite' in a series of articles published between 1854 and 1865.^{1–3} In these, Verneuil also suggested an association between hidradenitis and sweat glands, which had been discovered by Purkinje in 1833. Not having done any histopathological studies himself Verneuil conceded that his conclusions were purely based on the characteristic

distribution of the condition and that further studies into the link between sweat glands and HS would be required.¹

It took many decades and numerous studies until it was eventually accepted that HS is an acneiform disorder, which begins with follicular occlusion, rather than an infection of sweat glands.^{4–7}

In his publications Verneuil described the duration of a typical HS lesion as about 2 weeks. Since this early work no other study has systematically focused on the natural history of the disease. Jemec *et al.* described the socio-economic

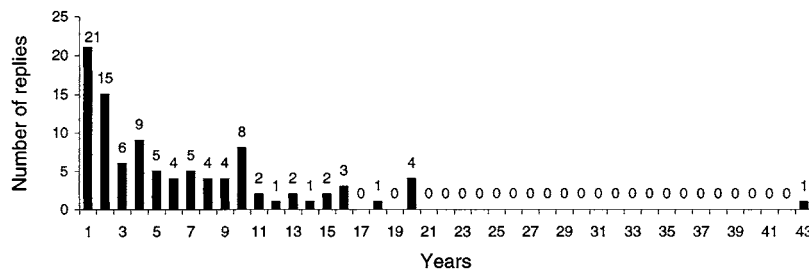


fig. 1 Replies to the question: How many years after it began was your condition at its worst?

characteristics of a large group of HS patients seen in a hospital setting, but did not address issues such as disease progression or average monthly incidence of inflammatory lesions.⁸ Such knowledge is important for the affected patients, but it is also important for physicians when they want to assess the effects of various interventions.

We have therefore conducted a questionnaire-based survey among HS patients from three hospitals in Nottinghamshire to find out more about the natural history of the condition.

Materials and methods

One hundred and fifty-six patients with a diagnosis of HS were identified through an electronic search of the diagnostic coding registers of the Departments of Dermatology and the Department of Plastic Surgery from three hospitals in Nottinghamshire. The records were traced back to 1993 in the two Nottingham hospitals and to 1995 in one hospital from Mansfield.

For patients derived from dermatology records a consultant dermatologist diagnosis was considered sufficient to qualify as a case. In those patients derived from the Department of Plastic Surgery a histological confirmation of the diagnosis was always available.

Patients were sent a questionnaire with 15 questions and a short introductory letter. The areas covered by the questions were: (i) basic demographic data; (ii) chronology of the disease in individuals, i.e. age of disease onset, timing of maximum disease activity, present disease activity; (iii) frequency and duration of boils; and (iv) factors associated with disease activity. Most patients also received a second questionnaire, consisting of the Dermatology Life Quality Index,⁹ the results of which will be presented elsewhere.

Results

Demographic data and chronology of disease

A total of 110 patients (70.5%) returned their questionnaires, 93 females and 17 males. The average age of our patients was 40.1 years (range 20–76 years). The average reported age of disease onset was 21.8 years (SD 9.8 years, range 5–54 years). In 12 patients who had not had any active disease for over a

year, the average reported age of disease onset was 34 years (SD 12.4 years, range 20–54 years; not statistically significant).

Only 23 patients were above 50 years old and in this subgroup males were much more common (nine males, 14 females) than in the overall study group. There was no female patient above the age of 65 years. Ninety-eight of the 110 responders still had experienced at least one boil within the past year. Patients had suffered an average disease duration of 18.8 years (SD 11.4 years, range 2–54 years) and the majority reported that their maximum disease activity had been in the early phases of their conditions (after a mean of 6.4 years, SD 6.4 years; fig. 1). Thirty-seven of 97 patients (38%) stated that they knew of at least one affected family member with HS.

Frequency and duration of boils

A broad range of replies was given to the question about the average number of painful boils suffered per month. Answers ranged from one painful lump per year to 30 per month. The median reply was two lesions per month (mean 4.6 lesions per month, SD 6.2). The average duration of a single painful boil was 6.9 days (SD 5.3), but 62% of responders also acknowledged the presence of at least one single tender lump that was permanently painful and failed to settle completely. In response to the question on whether 'boils' tended to settle with or without bursting 69% of responders said that more than half of their boils ultimately ruptured, and only 31% felt that the majority of their lesions remained 'blind', i.e. failed to burst.

Factors associated with disease activity

Patients listed a number of factors that could aggravate or relieve the intensity of their condition (Table 1). Heat, sweating and stress were the mostly commonly cited factors to exacerbate the disease, with 32% of responders observing a deterioration of their condition in the summertime. In 44% of females, menstruation reportedly led to an aggravation of the disease.

Discussion

Our study suggests that HS patients seen in the secondary care sector develop new boils at a rate of about two per month and

Table 1 Number and percentage of replies reporting factors which aggravate or relieve hidradenitis ($n = 97$, number of replies >97 as multiple responses were accepted in this open section of the questionnaire)

	Number of replies (%)
Worsening factors	
1. Sweating, heat, exercise	44 (45%)
2. Stress, fatigue, run down, pressure, low self-esteem	34 (35%)
3. Tight clothing, friction	16 (16%)
4. Deodorants, cosmetics, scented toiletries, shaving	13 (13%)
5. Weight	6 (6%)
6. Periods	5 (5%)
7. Others	9 (11%)
8. don't know	14 (14%)
Relieving factors	
1. Medical treatments	48 (49%)
various medicinal remedies	19 (19%)
surgery/lancing/laser/bursting	18 (18%)
antibiotics	11 (11%)
2. Life-style measures	39 (40%)
bath/shower/swimming	15 (15%)
loose/cotton clothing/cleanliness/drying/cold	11 (11%)
relaxing/no stress/sunshine/fresh air	9 (9%)
weight loss	2 (2%)
3. Pregnancy	2 (2%)
4. Nothing/don't know	23 (24%)

that individual boils take an average of about 1 week to resolve. These figures may in the future be helpful for sample size calculations of treatment studies for this condition.

In interpreting the results of this study it is important to appreciate that they were derived from a patient population from the secondary care sector and that they can thus not be assumed to be representative of all HS patients. They do however, represent a typical subset of severe cases that come under the care of dermatologists and plastic surgeons.

The high number of patients with a positive family history of the disease impressed us. Several authors have made the observation of a familial occurrence of HS in the past.^{10,11} In 1985 Fitzsimmons and Guilbert published pedigrees of a number of families in whom they noted autosomal dominant inheritance of HS.¹² In a recent study we were able to confirm their findings,¹³ which suggests a significant genetic contribution to the pathogenesis of HS in a proportion of patients.

The validity of our data is supported by a good response rate (70.5%) to our survey and patient demographics, which are compatible with those from other published studies. The large predominance of females among responders reflects previous observations that the condition is more frequent among women¹⁴ and would still persist if all the non-responders were male.

Our observation that the proportion of females was significantly lower in those patients aged 50 years or above (when compared against the overall study group) supports the notion that the disease tends to ease or subside in women after the

menopause.¹⁵ This is further supported by the absence of female patients over the age of 65 years in our study group.

We found an average age of disease onset of 21.8 years, which corresponds almost to the year with figures published by Jemec (average age of onset 23 years)¹⁶ and Harrison and Hughes (average age of onset 24.9 years).¹⁷ However, we found a higher average age of disease onset, albeit statistically not significant, among the small group of patients in whom the condition seemed to have subsided. This might suggest that patients with a late onset of HS have an overall better prognosis. Clearly, rate of onset will depend on the age of the examined population and it is possible that a study of older index cases will yield a later average age of disease onset.

In general, HS is a chronic disease as underlined by the finding that 90% of our patients still had active disease within the past year despite an average disease duration of nearly 19 years. Whether it is relentlessly progressive, as suggested by Hurley,¹⁸ remains unclear. The replies in our survey suggest that the majority of patients perceive the early phase of the disease as the worst part of it. This contradicts to a degree Hurley's statement, although recall bias and elements of adaptation to the disease process may partially explain our results. The impression of a relentlessly progressive disorder may be partially explained by our finding that almost two-thirds of patients acknowledged the existence of persistently painful boils that failed to settle. It appears possible that patients develop new boils at an unchanged rate throughout the course of their disease, some of which will eventually fail to subside in the usual manner and become chronic.

Disease flare-ups seem to occur particularly as a result of sweating, heat, stress and tight clothing or friction. The adverse effects from sweating and heat help to explain why nearly one-third of responders reported a deterioration of their condition in the summertime. Although only few women listed menstruation as a disease-aggravating factor in the open section of the questionnaire, direct questioning about this factor showed that almost half of all females reported perimenstrual exacerbations of their HS. Similar figures have been recorded by other authors.^{12,19}

Several patients were able to obtain some relief from their condition through certain life-style measures, particularly swimming, bathing and avoidance of tight-fitting clothing. Whereas some of this has been suggested before²⁰ it is not regularly included among the suggested treatments in most textbooks or patients' information leaflets.

Nearly a quarter of our patients had been unable to list any measure that helped their condition, despite an average disease duration of nearly 19 years. This is an indictment that the available treatments for HS are on the whole still unsatisfactory, including surgical approaches, with which almost half of our patients had previously been treated.

The textbook recommendation for treatment is systemic antibiotics,¹⁴ despite the fact that a poor response to standard

antibiotics has been so well documented that Mortimer made it a defining criterion for the definition of HS.²¹ The belief in the efficacy of antibiotics originates from the assumption that HS is an infection of the apocrine glands.^{14,20} It is now thought that HS is an acneiform disease in which pathogens are likely to be only secondary invaders of an as yet incompletely understood primary disease process.⁵ Still, approximately 10% of our patients reported some benefit from the use of systemic antibiotics. An explanation for this may come from our findings on the average duration of boils in HS. The observed figure of 6.9 days almost equals the average length of a course of antibiotics. It may thus not be the antibiotics but simply the natural history of the disease that has led to the registered improvement. No placebo-controlled study of the use of systemic antibiotics in HS has ever been published. Such a study seems urgently required to clarify this issue.

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